Posterior Cortical Atrophy
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Neurodegenerative condition characterized by progressive, fairly selective decline in visual-processing skills and other functions dependent on parietal, occipital and occipitotemporal regions of the brain.

Early age of onset; mid-50’s to early 60’s
Neuropathology

- Alzheimer’s most common underlying cause

- Other associated causes:
  - corticobasal degeneration
  - Dementia with Lewy Bodies
  - Prion Disease (CJD, familial fatal insomnia)
  - Subcortical gliosis
Clinical Symptoms

- **Visual symptoms**
  - Difficulty reading lines of text
  - Difficulty judging distances (minor car accidents, difficulty parking)
  - Difficulty identifying static objects in visual field
  - Difficulty with stairs, escalators

- Often first presents to ophthalmologist to rule out primary ocular disease
Neuropsychiatric Features

- Visuospatial and visuoperceptual impairments, alexia
- Balint’s syndrome
  - Simultanagnosia
  - Oculomotor apraxia
  - Optic ataxia
  - Environmental agnosia
- Gerstmann’s syndrome
  - Acalculia
  - Agraphia
  - Finger agnosia
  - Left-Right disorientation
- Deficits in working memory and limb apraxia
- Progression to involve anterograde memory, executive functions and linguistic skills
Example Visual Dysfunction

Healthy Individuals

Posterior Cortical Atrophy
Visuospatial Dysfunction (MMSE 26/30)
Associated Features

- Motor involvement
  - Extrapyramidal signs (41%)
  - Myoclonus (24%)
  - Grasp Reflex (26%)
Neuroimaging

- MRI
  - Cross-sectional voxel-based morphometry
    - Most significant reductions in occipital and parietal lobes followed by temporal lobe
    - Less left medial temporal and hippocampal atrophy
      - Typically atrophy by 5 years symptom duration atrophy is widespread
  
- FDG-PET
  - Structural changes in parieto-occipital regions

- PiB-PET
  - Increased accumulation of amyloid in occipital, parietal lobes; others show diffuse accumulation throughout all cortices
Figure 1. Serial MR images of a patient with posterior cortical atrophy (sagittal T1, axial FLAIR, coronal T1). Note the occipitoparietal atrophy (large white arrows) and the relative preservation of the hippocampus (small black arrow).
Figure 2: Neuropsychological and neuroimaging evidence of posterior cortical atrophy
Core features:

- Insidious onset and gradual progression
- Visual deficits in absence of ocular disease
- Relatively preserved episodic memory
- Verbal fluency and personal insight
- Symptoms including:
  - Visual agnosia, simultanagnosia, optic ataxia, ocular apraxia, dyspraxia and environment disorientation
- Absence of stroke or tumour
- Onset before 65 years
- Neuroimaging evidence of PCA or hypoperfusion
Management

- Memory
  - Acetylcholinesterase inhibitors
    - No trials, but often used with few case reports of benefit.

- Mood
  - Anti-depressants

- Social supports
  - Insight and memory preservation early

- Visual Impairment
  - Occupational therapy assessment and resources
  - May be legally blind – financial resources

